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# Multivisceral Transplant is a Viable Treatment Option for Patients with Nonresectable Intraabdominal Fibromatosis

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**Abbreviations**

**FAP**, familial adenomatous polyposis

**MVT**, multivisceral transplant

**Abstract**

***Background*** Intraabdominal fibromatosis often involves the mesentery root which is nonresectable by conventional surgery. Multivisceral transplant (MVT), as a potential cure to nonresectable fibromatosis, has rarely been reported and the prognosis is unknown. ***Methods*** Six patients who underwent MVT for intraabdominal fibromatosis were reviewed. Clinicopathological features, immunohistochemistry for β-catenin, p53, and Ki67, and outcomes were evaluated. Appropriate data for comparative analysis were obtained from a cohort of 24 patients who underwent conventional resection for intraabdominal fibromatosis. ***Results*** Among 6 MVT patients, 4 had familial adenomatous polyposis (FAP). Two patients had an initial intestinal

transplantation, three had multiple prior surgeries and two had adjuvant therapy. One patient died of hemorrhagic stroke shortly after MVT, and 5 patients (83%) survived with a median followup of 64 months. The 1-year and 5-year survival rates were 67% for all 5 patients. Two patients had recurrences after MVT and one of them had FAP. In comparison, 6 out of 24 patients who underwent conventional surgery had FAP; 6 (25%) had recurrences and 3 had FAP. For FAP patients, the mean recurrence time was 13 months for MVT versus 6 months for conventional surgery. Ki67 proliferative index, β-catenin, and p53 expression did not significantly correlate to recurrence. ***Conclusions*** MVT is a viable option for patients who have nonresectable intraabdominal fibromatosis with promising surviving rates, although recurrence still occurs. Surgical margin, Ki67 proliferative index, β-catenin, and p53 expression are not predicative for recurrence of fibromatosis.

## INTRODUCTION

Fibromatosis, first described by Stout et al., is a well-differentiated fibroblastic tumor. Morphologically, the tumor cells display features of infiltrating growth pattern, abundant collagen, low mitotic activity, local aggressiveness, and lack of malignant histology or potential for distant metastasis.[1] The tumor cells are positive for smooth muscle actin, desmin, calponin, and estrogen receptor-β, and negative for CD34.[2] At molecular levels, sporadic fibromatosis harbors mutations of gene *CTNNB1*, which encodes β-catenin protein. Therefore, the presence of nuclear expression of β-catenin is frequently used as a diagnostic tool.[3,4]

Based on locations, fibromatosis is further classified into two main types, superficial and deep. The former tends to involve superficial fascia or aponeurosis, and the deep type is categorized as extraabdominal (60%), abdominal wall (25%), and intraabdominal (desmoid tumor, 15%).[5] Multidisciplinary treatment, including surgery, radiation and adjuvant therapy, has been applied. Among them, surgery is the essential approach, although limitation exists[6]. First, intraabdominal fibromatosis, especially in patients with familial adenomatous polyposis (FAP), often involves the bowel or mesentery root, which is nonresectable by traditional surgery.[7] Secondly, wide resection with a negative surgical margin cannot always be achieved, which yields a high rate of recurrence.[7] Thirdly, repeated surgeries inevitably lead to formidable short gut syndrome that significantly impairs quality of life. All these post a challenge to conventional surgery.

Multivisceral transplant (MVT), as the last straw, is a potential treatment for patients with nonresectable fibromatosis that is beyond conventional surgery. Rare MVT cases have been reported, although the prognosis and predicators are largely unknown.[8-13] This report aimed to study the survival and recurrence in a series of 6 patients who had conventionally nonresectable fibromatosis and underwent MVT.

## MATERIALS AND METHODS

### *Patients*

Six patients who underwent MVT for nonresectable intraabdominal fibromatosis were identified in the Indiana University Health surgical pathology database from 2005 to 2015. The study was approved by the Institutional Review Board. Organ procurement, preservation, and the procedure of MVT were followed as previously described.[14,15] The evisceration consisted of dissection of the intraabdominal organs

including celiac and superior mesenteric artery inflow, liver, pancreaticoduodenal complex, small bowel, and part of colon. Lymph nodes in the retroperitoneum were also resected during the procedure. The cluster graft organs included stomach, small bowel, and portion of colon, liver, and pancreas. All patients received antibody-based induction immunosuppression with calcineurin-inhibitor–based maintenance and low dose steroids, as well as protocols of graft surveillance and infectious prophylaxis.[16,17]

In comparison, 24 patients who underwent conventional surgeries for intraabdominal fibromatosis were identified from the same time period. Patients who had abdominal wall and parietal fibromatosis were excluded from the study.

***Histomorphologic review***

The cases were reviewed by two pathologists (ZC and JL) to compare the histomorphologic features. Tumor site, size and number, surgical margin status, involved organs, tumor necrosis, and mitosis were evaluated. Mitosis was assessed at a magnification of ×400 using an Olympus BX51 microscope. Representative fields were selected and at least 500 nucleated tumor cells were assessed for each case.

***Immunohistochemical evaluation***

Immunohistochemistry for assessing Ki67, p53, and β-catenin was performed on the tumor tissue of all MVT cases. A representative formalin-fixed, paraffin-embedded tumor section was used. Briefly, deparaffinized tissue sections were stained with antibodies against Ki67, p53, or β-catenin. Immunohistochemical staining was carried out with a mouse monoclonal anti-Ki67 (dilution 1:1; DAKO, Carpinteria, CA),

p53 (dilution 1:1, ONCOGENE SCIENCE, Cambridge, MA), and β-catenin (dilution 1:1, ZYMED, South San Francisco, CA). A high pH buffer solution in a “PT module” was used for antigen retrieval followed by incubation times of 10 minutes each with primary antibody, Envision FLEX+M linker, Envision FLEX/horseradish peroxidase (Dako, Carpinteria CA), and diaminobenzidine..

Expression of Ki67 proliferation index and p53 was calculated by manual counting of camera-captured/printed image[18] of at least 50 HPFs and/or 2000 nucleated tumor cells (40X objective) in the area of expression and reported as a number of positive cells per 10 HPFs using an Olympus BX51 microscope.

### *Statistics*

Categorical data were compared using the $\chi 2$ test. Continuous data were compared using Student *t* test. A *P* value less than 0.05 is considered statistically significant.

## RESULTS

### *Demographics*

Three patients who received MVT were men (50%) and the average age of MVT group was 41 years (range, 29–60 years). Eleven patients who received conventional surgery were men (46%) and the average age was 42 years (range, 15–70 years). There was no age or gender difference in two study groups ($P > 0.05$).

Four patients in MVT group and 6 patients in conventional group had FAP. Two FAP patients in MVT group had an initial intestine-only transplantation before MVT, and the allografts failed. Three patients in MVT group had prior multiple small bowel resections or subtotal colectomy. These patients in MVT group presented with

short gut syndrome or frozen abdomen (Table 2). Two MVT patients (#1 and 4) had received adjuvant therapy (tamoxifen, methotrexate, or imatinib, Table 2).

***Histopathologic evaluation***

As shown in Table 1, the primary site was mesentery for all patients in the study. The average size of tumor was 14.6 cm (range, 4.5–25 cm) in MVT group compared to that of 8.5 cm (range, 1.5–26 cm) in conventional group ($P$=0.19). The average number of tumor in MVT group was 3 (range, 1–9) that was statistically higher than that of 1.4 (range, 1–4) in conventional group ($P$=0.0005). Three patients in MVT group had positive resection margins (50%) compared to that of 54% in conventional group. Involved organs in MVT group included small bowel (100%), stomach (33%), pancreas (33%), spleen (33%), and liver (16%) which were more extensive in contrast to those of small bowel (100%), stomach (4%), and colon (12%) in conventional group. Necrosis was not present and mitotic figures were rare in both study groups. The clinicopathological features of each individual patient who had MVT were shown in Table 2.

***Prognosis***

As shown in Table 3, among 6 MVT patients, 1 died of hemorrhagic stroke shortly after MVT and all patients in conventional group survived the procedure. For the 5 patients who survived MVT, median followup time was 64 months (range, 4–116 months) compared with 49 months (range, 1–106 months) in conventional group. One patient died 4 months post-MVT due to sepsis, and 1 patient in conventional group passed away after 6 months due to gastrointestinal tract bleeding. The overall 1-year and 5-year survival rates were both 67% in MVT group compared to those of

96% in the conventional group. In MVT group, 2 patients (40%) had recurrences with a median recurrence time of 9 months (range, 4–13 months) in contrast to 6 patients (25%) in the conventional group with a median recurrence time of 23 months (range, 5–74 months).

One FAP patient passed away 4 months post-MVT due to sepsis in contrast to none in conventional group. The 1-year and 5-year survival rates for FAP patients in MVT group were 75% compared with 100% in conventional group. One FAP patient (25%) in MVT group had recurrence 13 months post-MVT and 3 (50%) FAP patients in control group had recurrences with a median time of 6 months (5, 6 and 33 months). The single non-FAP patient in MVT group had a recurrence time at 4 months after MVT. Among 18 non-FAP patients in conventional group, 3 (17%) had recurrence with a median time of 32 months (14, 32 and74 months)).

Further analysis was performed in 5 MVT patients respective to recurrence. As shown in Table 4, recurrence occurred in 2 of 3 patients whose surgical margins were positive. Although there was likely a trend that positive margin was associated with recurrence, it was not statistically significant ($P > 0.05$).

***Immunohistochemistry study***

Immunohistochemistry for Ki67, p53, and β-catenin was performed on the tumor tissue in MVT group (Figure 1; Table 4). In the 2 patients with recurrent disease, 83% (mean; range, 82–84%) of tumor cells were positive for nuclear β-catenin immunostaining compared to 51% (mean; range, 31–64%) in 3 nonrecurrent cases ($P>0.05$). Ki67 proliferation indexes were 4% (mean; range, 3–5%) in recurrent cases compared with 2% (mean; range, 0–3%) in nonrecurrent group ($P>0.05$). p53 immunoreactivity was 5% (mean; range, 2–8%) versus 4% (mean; range, 0–8%)

between recurrences and nonrecurrences with no statistical significance (*P*>0.05). Although the expression levels of Ki67, p53, and β-catenin in recurrent tumors appeared higher than those of the nonrecurrence, there was no statistical significance.

## DISCUSSION

MVT was first performed 30 years ago by Dr. Starzl et al. in a child with short gut syndrome and secondary liver failure.[19] Since then, MVT has been performed around the world with varying degrees of success. Major indications for MVT in both children and adults include gastroschisis, necrotizing enterocolitis, intestinal atresia, mesenteric thrombosis, trauma, and intestinal dysmotility syndrome.[20] Recently, nonresectable intraabdominal fibromatosis involving the mesenteric root has been indicated for MVT. However, given the complex natures of MVT and associated high risks, only a handful studies have been published (Table 5).[8-13] The first MVT for intraabdominal fibromatosis was reported by Misiakos et al. in 1999.[8] The patient had Gardner syndrome and fibromatosis with multiple episodes of intestine resections and eventually developed short gut syndrome. Additionally, prolonged total parenteral nutrition impaired the liver. After MVT, fibromatosis recurred in this patient in thoracic wall twice that was successfully resected. In 2001, Chatzipetrou et al. reported two MVT cases, in which only one patient survived the procedure without recurrence.[9] In 2002 and 2005, Jovine et al. and Tryphonopoulos et al. published 3 and 2 cases, respectively, with a total of 4 surviving patients, although the status of recurrences was not provided.[10,11] In Tryphonopoulos' study, 3 patients had intestinal autotransplantation and all survived the procedure without recurrence of the disease.[11] In 2011, Cruz et al. reported 10 liver-sparing modified MVT.[12] All patients

had FAP and short gut syndrome with extensive fibromatosis involving the pancreas and duodenum. All patients survived the procedure without recurrence. The overall cumulative survival rate was 90% at 1 year and 77% at 10 years. In 2013, Nikeghbalian et al. published one case. The patient survived the procedure without recurrence.[13] In the current report, 6 patients received MVT for nonresectable fibromatosis with a median followup of 64 months. To our knowledge, this represents the second largest series in the literature.

The overall 1-year and 5-year survival rates were both 67% for MVT patients; and the recurrence rate was 40% for the 5 followed-up patients. In summary, 22 (88%) out of 25 patients who survived MVT have been reported so far in the English literature. Among them, 3 (15%) had known recurrence. In comparison, the overall 1-year and 5-year survival rates were both 96% for the conventional surgery group, and the recurrence rate was 25% for the followed-up patient. These results are comparable with the 1- and 5-year recurrence-free survival rates of 81.3% and 52.8 % reported in Johns Hopkins study.[21]

The 6 patients in MVT group had multiple tumors and the tumor load was larger extensively involving multiple visceral organs in comparison to those in conventional group. Among 6 MVT patients, most had prior surgeries including multiple small bowel resection (1), subtotal colectomy (2), and isolated small intestinal transplantation (2). Among them, two patients had adjuvant therapy. But all these treatments failed to control the tumor progression which fatally involved the mesentery root and multiple visceral organs. As noted, the largest size of the tumor in one of our patient (#3) was less than 5 cm, but this patient had 4 fibromatosis tumors (4.5 cm, 4 cm, 3.5 cm and 2.5 cm) involving multiple visceral organs (liver, stomach, pancreas, small bowel and spleen) that caused short gut syndrome.

Although autologous reconstruction of superior mesentery artery base, auto-transplantation or ex vivo resection would be feasible for some small-size tumors, they were not suitable in this particular case, because of the extensive tumor involvement in multiple important visceral organs that made it nonresectable by conventional surgery. It is worth mentioning that MVT is truly the last straw that should be considered for fibromatosis patient when all the other possible options are excluded, given the complex nature and the high risk of the procedure.

An interesting finding of our study is that FAP patients in MVT group had a trend of lower recurrence rate compared to their counterparts in conventional surgery, which keeps in line with Cruz's observation.[12] Although sporadic and FAP-associated fibromatosis both have enhanced β-catenin protein expression, the mechanisms are different. It is known that the characteristic *APC* mutation in FAP causes nuclear accumulation of β-catenin protein, while the *CTNNB1* mutation leads to nuclear accumulation of β-catenin protein in sporadic fibromatosis.[22] This difference in pathogenesis might play a role in recurrence, although further investigation is warranted.

Prognostic factors for recurrence in MVT were also studied. Our data indicate that surgical margin, Ki67 proliferative index, β-catenin, and p53 expression do not significantly correlate to recurrence. Given the small size of cases, more studies from multiple transplant centers are warranted to finalize the conclusion.

In summary, MVT is a viable option for patients with advanced intraabdominal fibromatosis who are, otherwise, ultimately fatal and not amenable to conventional options. Surgical margin, Ki67 proliferative index, β-catenin, and p53 expression are not predicative for recurrence of fibromatosis in patients who takes MVT.

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Table 1. Clinical and histopathologic comparison of intraabdominal fibromatosis in patients with multivisceral transplant (MVT) or conventional surgery.

| | MVT (n=6) | Conventional surgery (n=24) |
|---|---|---|
| Primary site | Mesentery | Mesentery |
| Mean size (range) | 14.6 cm (4.5–25) | 8.5 cm (1.5–26) |
| Average number of tumor (range) | 3 (1–9) | 1*(1–4) |
| Margin positivity | 3 (50%) | 13 (54%) |
| Organs involved | Small bowel (100%)<br>Stomach (33%)<br>Pancreas (33%)<br>Spleen (33%)<br>Liver (16%) | Small bowel (92%)<br>Stomach (4%)<br>Colon (12%) |
| Necrosis | Not present | Not present |
| Mitosis | Rare | Rare |

*$P$ =0.0005 compared to the MVT group.

Table 2. The clinicopathological characteristics of the 6 patients who had multivisceral transplant for intraabdominal fibromatosis.

| Patient | 1 | 2 | 3 | 4 | 5 | 6 |
|---|---|---|---|---|---|---|
| Age/Sex | 33/M | 60/F | 29/M | 33/F | 50/F | 39/M |
| Number of tumor | 1 | 3 | 4 | 9 | 3 | 1 |
| Size of tumor (the largest) | 15 cm | 15 cm | 4.5 cm | 16 cm | 25 cm | 12 cm |
| FAP status | Yes | Yes | Yes | No | No | Yes |
| Organ involvement | Small bowel | Small bowel | Liver, stomach, pancreas, small bowel, spleen | Stomach, Small bowel, pancreas, abdominal wall, | Small bowel, spleen | Small bowel |
| On TPN | No | No | Yes | No | Yes | Yes |
| Liver failure | No | No | No | No | No | No |
| Organs transplanted | Stomach, small bowel, portion of colon, liver, and pancreas | Stomach, small bowel, portion of colon, and pancreas | Stomach, small bowel, portion of colon, liver, and pancreas | Stomach, small bowel, portion of colon, and pancreas | Stomach, small bowel, portion of colon, liver, and pancreas | Stomach, small bowel, portion of colon, liver, and pancreas |
| Necrosis | Not present | Not present | Not present | Not present | Not present | Not present |
| Ki67 | 0.0% | 5.7% | 5.7% | 2.8% | 3.1% | 0.6% |
| MVT complication (infection, shock) | No | No | No | No | Died of peri-operative hemorrhagic stroke | Died of sepsis 4 m later |
| Pre-MVT condition and pre-MVT surgery | Subtotal colectomy, Short gut syndrome | Frozen abdomen | Multiple small bowel resections, short gut syndrome | Omental and subtotal colectomy; short gut syndrome | Initially pre-MVT intestinal transplantation and allograft failure | Initially pre-MVT intestinal transplantation and allograft failure |
| Adjuvant therapy (chemo or tamoxifen) | Yes(Tamoxifen and methotrexate) | No | No | Yes (Imatinib and tamoxifen) | No | No |
| Follow up time | 116 | 64 | 5 | 78 | 0 | 4 |

| | | | | | | |
|---|---|---|---|---|---|---|
| (months) | | | | | | |
| Recurrence and time elapsed (months) | No | No | Yes (4) | Yes (13) | n/a | No |
| Prognosis (dead/alive) | Alive | Alive | Alive | Alive | Dead | Dead |

Abbreviation: FAP, familial adenomatous polyposis; MVT, multivisceral transplant; TPN, total parenteral nutrition.

Table 3. Followup and recurrence of intraabdominal fibromatosis.

| | Total | | FAP | | Non-FAP | |
|---|---|---|---|---|---|---|
| | MVT* | Conventional | MVT | Conventional | MVT | Conventional |
| Number | 5 | 24 | 4 | 6 | 1 | 18 |
| Followup time (months; range) | 64 (4–116) | 49 (1–106) | 71 (4–116) | 46 (6–106) | 5 (5) | 49 (1–98) |
| Recurrence | 2 (40%) | 6 (25%) | 1 (25%) | 3 (50%) | 1 (100%) | 3 (17%) |
| Medium recurrence time (months; range) | 9 (4–13) | 23 (5–74) | 13 (13) | 6 (5–32) | 4 (4) | 32 (14–74) |

*One patient died shortly after MVT due to hemorrhagic stroke.

Abbreviation: FAP, familial adenomatous polyposis; MVT, multivisceral transplant

Table 4. Recurrence of intraabdominal fibromatosis in multivisceral transplant (MVT) patients

| MVT | Recurrent | Nonrecurrent |
|---|---|---|
| Number of patients* | 2 | 3 |
| Followup period (months; range) | 42 (5–78) | 64 (4–116) |
| Recurrence time (months; range) | 9 (4–13) | N/A |
| β-catenin-nuclear positive immunoreactivity (range) | 83% (82–84%) | 58% (31–64%) |
| Ki67-positive immunoreactivity (range) | 4% (3–5%) | 1% (0–3%) |
| p53-positive immunoreactivity (range) | 5% (2–8%) | 3% (0–8%) |
| Positive surgical margin | 2 (100%) | 1 (33%) |

*One patient died shortly after MVT due to hemorrhagic stroke and this case is excluded from the analysis of recurrence.

Table 5. Published series of multivisceral transplant to treat nonresectable intraabdominal fibromatosis

| Reports | Year | Case number | Survived MVT | Recurrence | Followup time (median months) |
|---|---|---|---|---|---|
| Misiakos[8] | 1999 | 1 | 1 | 1 | N/A |
| Chatzipetrou[9] | 2001 | 2 | 1 | 0 | 43 (range 38–48) |
| Jovine[10] | 2002 | 2 | 2 | N/A | 6 (range 5–6) |
| Tryphonopoulos[11] | 2005 | 3 | 2 | N/A | 63 (range 13–108) |
| Cruz[12] | 2011 | 10 | 10 | 0 | 50 (range, 18–128) |
| Nikeghbalian[13] | 2013 | 1 | 1 | 0 | 31 |
| Chi (current) | 2017 | 6 | 5 | 2 | 64 (range, 4–116) |
| Summary | N/A | 25 | 22 (88%) | 3 (15%) | 4–128 |

Abbreviation: MVT, multivisceral transplant; N/A, not applicable.

**Figure Legend**

**Figure 1.** Intraabdominal fibromatosis. **(A)** High-power view of fibromatosis showing cytologically bland spindle- or stellate-shaped fibroblastic cells arranged into intersecting fascicles with collagenous deposition (H&E, magnification x200). **(B)** Immunohistochemically, fibroblastic cells are strongly positive for β-catenin in a nuclear staining pattern (magnification x200). Immunostaining for Ki67 proliferative index **(C)** and p53 **(D)** highlights rare spindle cells (magnification ×200).